

Classification of ventricular septal defects*

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SUMMARY A classification with clinical significance is proposed for ventricular septal defect based on the study of 220 hearts with defects of the ventricular septum. All had atrioventricular and ventriculoarterial concordance with normal relations of cardiac structure. For the purpose of classification, the ventricular septum was considered as possessing muscular and membranous portions, the muscular septum itself being divided into inlet, trabecular, and outlet (or infundibular) components. Defects were observed in the area of the membranous septum, termed perimembranous defects; within the muscular septum, termed muscular defects; or in the area of septum subjacent to the arterial valves, termed subarterial infundibular defects. Perimembranous defects were found extending either into the inlet, trabecular, or infundibular septa. Muscular defects were found in or between the inlet septum, trabecular septum, or infundibular septum. Review of the angiograms showed that the classification was easy to use in the catheterisation laboratory, and our observations suggest that the precision thus obtained has considerable surgical significance.

In this report, we present a simplified concept for the classification of ventricular septal defects based on the study of over 200 pathological specimens with atrioventricular concordance, ventriculoarterial concordance, and usual relations of intracardiac structures. We have shown it to be useful for angiographic diagnosis and we believe it to have considerable surgical relevance.

Subjects and methods

The hearts studied were taken from the cardio-pathological collections of the Cardiothoracic Institute, Brompton Hospital, London; the Royal Liverpool Children's Hospital; the University of Sheffield; Grimsby General Hospital; Welhelmina Gasthuis, Amsterdam; St Antonius Ziekenhuis, Utrecht; Department of Anatomy, Rijksuniversiteit, Leiden; the Mayo Clinic, Rochester, Minnesota; and the Children's Hospital of Pittsburgh, Penn-

sylvania. In all, 220 hearts were studied (Table). Only hearts with isolated or multiple ventricular septal defects in the presence of atrioventricular and ventriculoarterial concordance were included. We excluded hearts with tetralogy of Fallot¹ but included hearts with ventricular septal defect associated with valvar pulmonary stenosis. Some of the hearts have previously been illustrated² and an earlier concept³ was based on study of others but the series as a whole has not been analysed previously.

The defects were studied with particular reference to their relation to the atrioventricular valves, the arterial valves, and the muscular bundles of the right ventricle, including the medial papillary muscle. The defects will be described as viewed by the angiographer or clinician, with the patient considered as being in an upright position. When the hearts have been photographed they have been oriented as far as possible to achieve this positioning. Photographs have been taken from the left ventricle, with the apex of the heart to the bottom of the frame and the aortic valve to the top.

The angiographic feasibility of the suggested classification was assessed by studying the angiograms from patients with ventricular septal defects

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and atrioventricular and ventriculoarterial concordance and usual relations investigated in the Department of Paediatric Cardiology, University of Alabama in Birmingham, Birmingham, Alabama (courtesy of Dr L M Barger, Jr). Illustrative examples of the classification were obtained from these patients.

Results

(1) CONSTITUENTS OF NORMAL VENTRICULAR SEPTUM AND TERMINOLOGY OF MUSCLE BUNDLES

For the purpose of classification of septal defects, we have considered the ventricular septum as having four components: the inlet septum, trabecular septum and outlet or infundibular septum (together making up the muscular septum), and the membranous septum (Fig. 1). The inlet septum separates the septal cusps of the mitral and tricuspid valves. It merges imperceptibly with the trabecular septum which is the largest part, extending out to the ventricular apices, and separates the finely trabeculated left ventricular apex from the more coarsely trabeculated right ventricular apical zone. It in turn merges imperceptibly with the outlet or infundibular septum which separates the right and left ventricular outlet tracts, being considerably more extensive on its right ventricular aspect than on the left. The membranous septum in the normal heart is a small structure, divided into two parts by the insertion of the septal leaflet of the tricuspid valve. This insertion produces an atrioventricular and an interventricular component, the sizes of which vary in different hearts.

In order to describe ventricular septal defects, it is also necessary to define the nature of muscular

trabeculae and bands observed in the normal right ventricle. The normal pulmonary and tricuspid valves are separated by an extensive ledge termed in the normal heart the crista supraventricularis. It has two components. Part of the normal crista additionally separates the aortic and pulmonary valves and is the outlet or infundibular septum. The other part of the crista between the pulmonary and tricuspid valves is the ventriculoinfundibular fold.⁴ The extensive septal trabeculation of the right ventricle is also considered to be part of the crista by some authors⁵ and is termed the 'septal band'. Though yet others do not consider the septal band to be part of the crista,⁶ in our opinion it is better considered a separate structure and named the trabecula septomarginalis.⁴ It is a septal trabecula on the right aspect of the trabecular septum, forming the superficial stratum of the septum. An important small papillary muscle of the tricuspid valve arises from the posterior limb of the trabecula septomarginalis and supports the antero-septal commissure of the tricuspid valve. It is the medial papillary muscle. The apical part of the trabecula septomarginalis is frequently enlarged and hypertrophied, and may form a 'two-chambered right ventricle'. This feature does not affect the classification of septal defects.

(2) CATEGORISATION OF VENTRICULAR SEPTAL DEFECTS (Table)

Our study has shown that on the basis of the septal division given above, isolated defects exist for the most part in the area of the membranous septum (Fig. 2). However, as indicated by Becu *et al.*⁵ and endorsed by others, these defects additionally involve the area of muscular septum surrounding the membranous septum itself, which may be present as

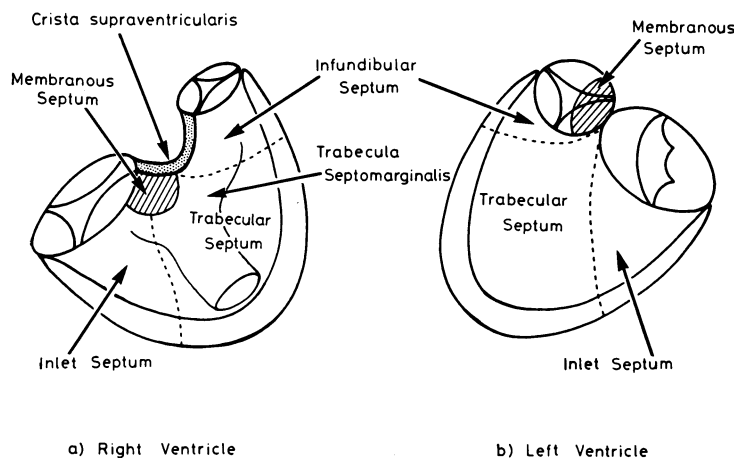
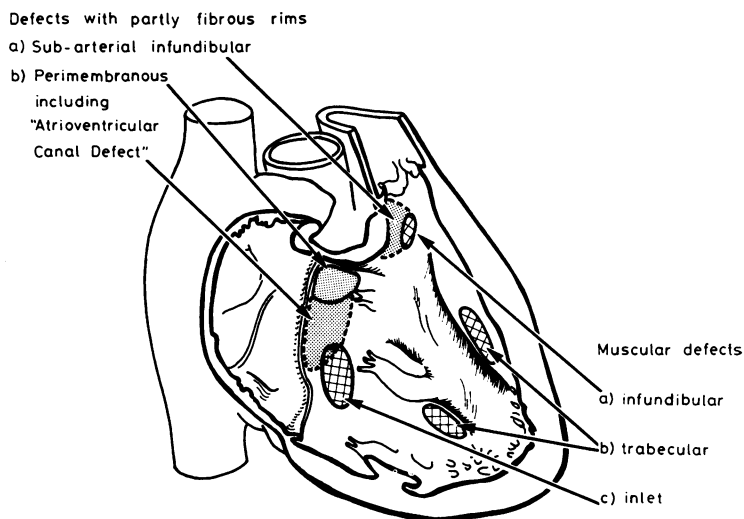


Fig. 1 Diagram illustrating the different muscular components of the ventricular septum and their conjunction with the membranous septum.

Fig. 2 Diagram illustrating the proposed classification of ventricular septal defects.



a remnant in the roof of such defects. Many have indicated that 'membranous' is an inappropriate term for such defects. We have taken note of these objections and have employed the term *perimembranous* to describe these defects. Perimembranous defect may involve the area of either the inlet, trabecular, or infundibular septum contiguous with the area normally closed by the membranous septum (Fig. 3).

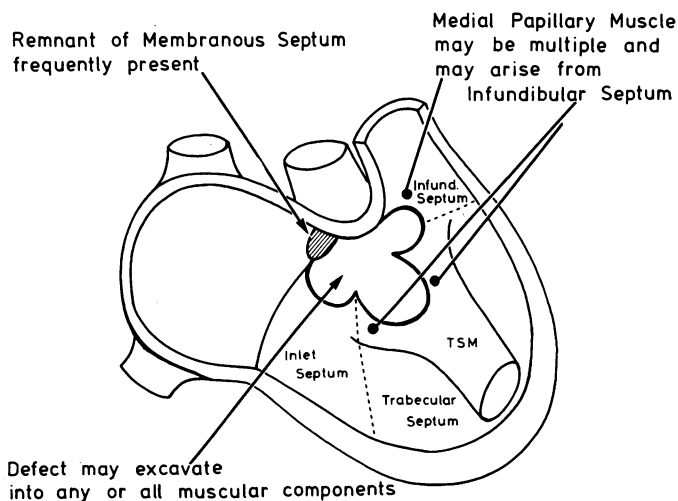
In contrast to perimembranous defects, in which part of the rim of the defect is always formed by part of the central fibrous body, other defects have purely muscular rims (Fig. 2). They are collectively termed *muscular defects*, and may exist in the area between

the inlet and trabecula septa (termed inlet muscular defects), within the trabecular septum itself (termed trabecular muscular defects), or between the in-

Table Categorisation of ventricular septal defects in 220 hearts

Type	Subcategories	No. studied
Perimembranous	(a) Inlet	55
	(b) Trabecular	56
	(c) Infundibular	42
Muscular	(a) Posterior (inlet)	15
	(b) Trabecular	40
	(c) Infundibular	3
Subarterial infundibular		12
Mixed defects		3

Fig. 3 Diagram illustrating the types of perimembranous septal defect. TSM, trabecula septomarginalis; Infund, infundibular.



PERIMEMBRANOUS VSD

fundibular and trabecular septa (termed infundibular muscular defects). A further type of defect exists in the area normally formed by the infundibular septum which does not have a purely muscular rim, and is therefore not a muscular defect, but equally is not a perimembranous defect. The non-muscular rim of these defects is formed by the contiguous arterial valves and they are termed *sub-arterial infundibular defects* (Fig.2).

I: Perimembranous defects

The unifying feature of these defects was that all had the area of tricuspid-mitral-aortic fibrous continuity (central fibrous body) as part of their rim (Fig. 3). The precise boundaries of the defects, and their relation to the aortic and atrioventricular valves, depended on whether the defect extended

primarily into the inlet septum, the trabecular septum, or the infundibular septum.

(a) *Defects extending into inlet septum* When viewed from the right ventricular aspect (Fig. 4A), these defects were beneath the septal cusp of the tricuspid valve. Their atrial margin was the area of tricuspid-mitral continuity. The floor and anterior margin of the defect was the crest of the inlet and trabecular septa which ran up in the roof to merge with the infundibular septum. This septum was normally positioned and fused with the trabecular septum as in the normal heart. The medial papillary muscle was usually found above the defect. From the left ventricular aspect it was seen that the posterior margin of the defect was an extensive area of aortic-mitral-tricuspid continuity. The non-coronary cusp was in extensive continuity with the

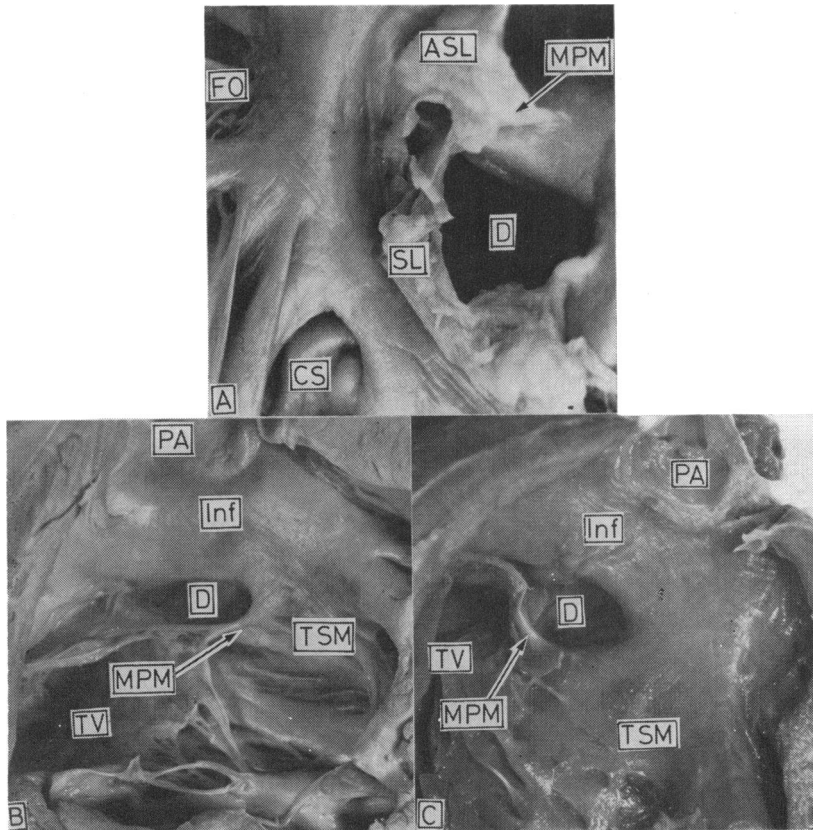


Fig. 4 Figures of perimembranous defects extending predominantly into the inlet septum (Fig. 4A); the trabecular septum (Fig. 4B) and the infundibular septum (Fig. 4C). The inlet defect is shown in close up. It lies beneath the septal leaflet (SL) of the tricuspid valve. Medial papillary muscle (MPM) and the anterosuperior leaflet (ASL) are above the defect. FO, fossa ovalis; CS, coronary sinus. The trabecular defect (D) is behind the tricuspid valve (TV) extending between the limbs of the trabecula septomarginalis. The infundibular defect has some degree of override so that the aortic cusps are visible.

tricuspid valve, making the aortic valve cusps the roof of much of the defect (Fig. 5A). In four hearts the septum was 'scooped' out up to the crux cordis as is usually found in atrioventricular defects (atrioventricular canal malformations or endocardial cushion defects) (Fig. 6) and in two of these hearts there was a cleft of the mitral valve. The roof of the defect in all these hearts was the infundibular septum which separated the right coronary aortic cusp from the edge of the defect. There was no malalignment of the septal structures in these hearts.

(b) *Defects extending into trabecular septum* In these specimens the defect was more elongated and extended towards the ventricular apex. When viewed from the right ventricle, its posterior rim

was beneath the septal leaflet of the tricuspid valve, and its atrial border was a small area of aortic-mitral-tricuspid continuity (Fig. 4B). Its floor was the inlet septum, merging anteriorly with the trabecular septum buttressed by the trabecula septomarginalis. The roof of the defect was the infundibular septum, which was normally aligned relative to the trabecular septum so that no aortic overriding was present (Fig. 4B). The medial papillary muscle tended to take origin from the midpoint of the right side of the defect, and was attached to the septal commissure of the tricuspid valve across the defect. This relation was better seen from the left (Fig. 5B), this view also showing well the primary extension into the trabecular septum. The long axis of the defect was from the aortic valve towards the left ventricular apex. In many hearts

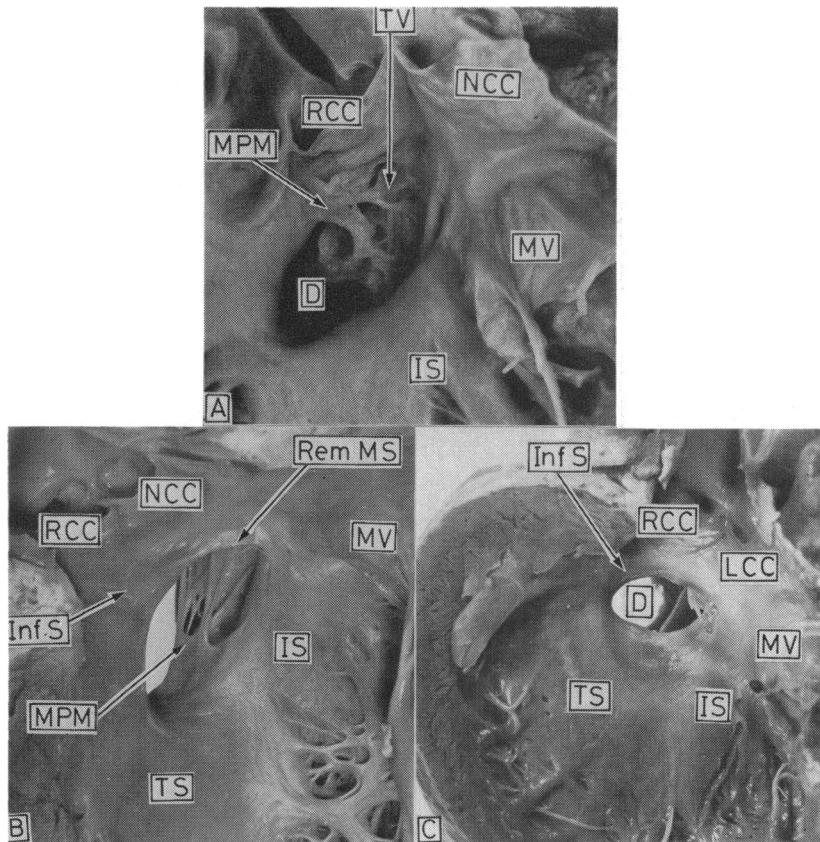


Fig. 5 The same three defects illustrated in Fig. 4 viewed from the left ventricular aspect. Fig. 5A shows the inlet defect. The inlet septum (IS) is deficient. RCC, right coronary cusp of aortic valve; NCC, non-coronary cusp; MV, mitral valve. Fig. 5B shows the trabecular defect. The inlet septum is better formed. The defect points toward the apex and a remnant of the membranous septum (Rem MS) separates the defect from the aortic valve. Inf S, infundibular septum. Note the different orientation of the infundibular defect shown in Fig. 5C.

remnants of tissue were delaminated from the central fibrous body in the roof of the defect, as viewed from the left. The aortic valve noncoronary cusp was in continuity with the mitral valve, but the membranous septal remnant separated the aortic valve from the tricuspid valve (Fig. 5B). The anterior margin of the roof of the defects when viewed from the left ventricle (Fig. 5B) was the infundibular septum, which formed a muscular bar separating the right coronary cusp from the defect. In these defects, therefore, a much smaller area of the aortic valve was directly contiguous with the defect as compared with the perimembranous inlet defect (compare Fig. 5A and 5B).

(c) *Defects extending into the infundibulum* The infundibular septum of these hearts was less well formed than in the other perimembranous defects

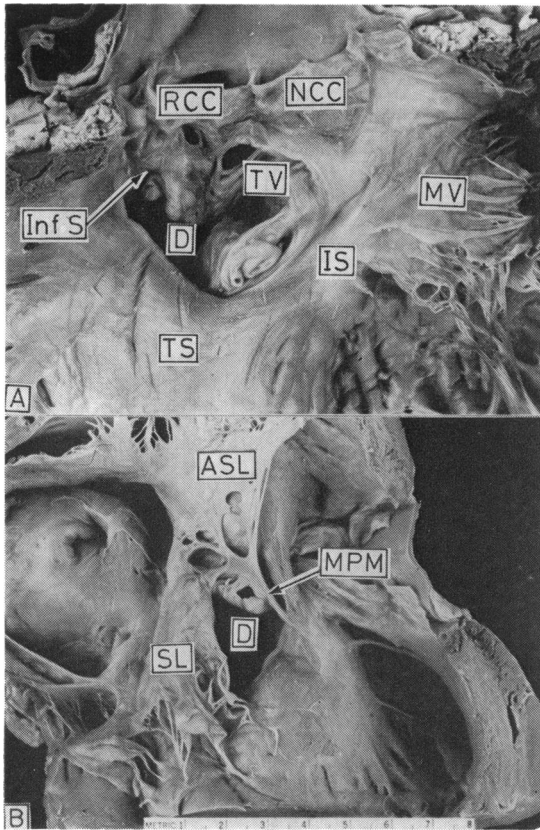


Fig. 6 Photograph of an inlet defect (of atrioventricular canal type). The inlet septum is deficient right to the crux of the heart. Fig. 6A shows the left ventricular aspect and 6B the right ventricular aspect. There was no cleft in the mitral valve (abbreviations as before).

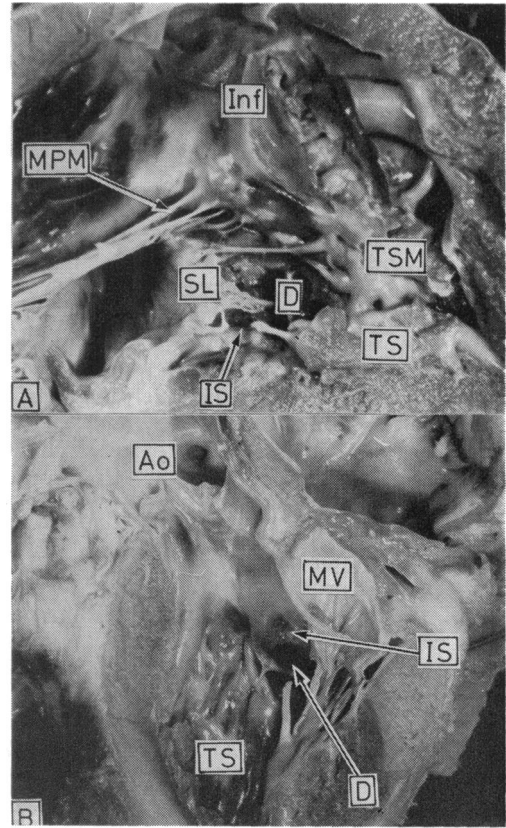


Fig. 7 Photographs illustrating a muscular defect between the inlet septum and the trabecular septum. Fig. 7A is a right ventricular view, 7B a left ventricular view (abbreviations as before).

and was usually raised to the right so that the aortic valve overrode the right ventricle (compare Fig. 4B and 4C). The atrial edge of the defect as viewed from the right ventricle was again an area of tricuspid-mitral continuity, and this became continuous anteriorly with the overriding aortic valve which formed the roof of the defect. The infundibular septum formed the anterior edge of the roof merging with the trabecular septum and trabecula septomarginalis in the anteroinferior rim of the defect. The medial papillary muscle was below the defect (Fig. 4C), but accessory papillary muscles were occasionally observed arising from the infundibular septum. The anterior extension of the defect was well seen from the left ventricular aspect (Fig. 5C). Its long axis, in contrast to the trabecular perimembranous defect, was oriented from left to right (compare Fig. 5B and 5C). The posterior margin of the defect, as viewed from the left

ventricle, was the central fibrous body. In the posterior margin of the roof, the aortic valve noncoronary cusp was directly contiguous with the defect as a consequence of the aortic overriding, but in the anterior margin the valve was separated from the defect by the infundibular septum (Fig. 5C).

II: Muscular defects

(a) *Defects in inlet area of muscular septum* These defects were beneath the septal cusp of the tricuspid valve (Fig. 7A), but possessed completely muscular rims. When viewed from the left ventricle, a rim of inlet septum was present beneath and between the septal cusps of the tricuspid and mitral valves (Fig. 7B). This rim crossed obliquely the crest of the trabecular septum. It extended from the right ventricle, where it was fused with the trabecula septomarginalis posteriorly into the left ventricle, where it fused with the trabecular septum. Frequently this latter fusion point was overlaid by a prominent posterior trabecula. The size of the defect varied according to the degree of development of the trabecular septum. A membranous septum was present and intact in these hearts, having both atrioventricular and interventricular components.

(b) *Defects in the trabecular area of the muscular septum* These defects in the muscular septum were frequently multiple and better appreciated from

the left than the right side. The openings as seen from the right ventricle were closely related to the edges of the trabecula septomarginalis (Fig. 8A). From the left it was seen that the defects were within the trabecular septum (Fig. 8B) but their position was variable. The membranous septum was normally formed in these hearts.

(c) *Defects in infundibular area of septum* Three specimens were encountered in which a defect with entirely muscular rims was located between the ventricular outflow tracts. As viewed from the right ventricle, the roof of the defect was the remnant of the infundibular septum. This merged anteriorly with the anterior limb of the trabecula septomarginalis (Fig. 9A) which formed the floor of the defect. Posterior, the infundibular septum merged with the ventriculoinfundibular fold and the posterior limb of the trabecula septomarginalis. This fusion of muscular structures formed a muscular rim in front of a normally formed and positioned membranous septum. When viewed from the left ventricle, the defect was seen to be completely separated from the aortic valve by the infundibular septum. Posterior to this the aortic valve was continuous with the central fibrous body (Fig. 9B). The difference between this defect and the perimembranous infundibular defect was that the aortic valve did not form part of the rim of the defect (compare Fig. 5C and 9B).

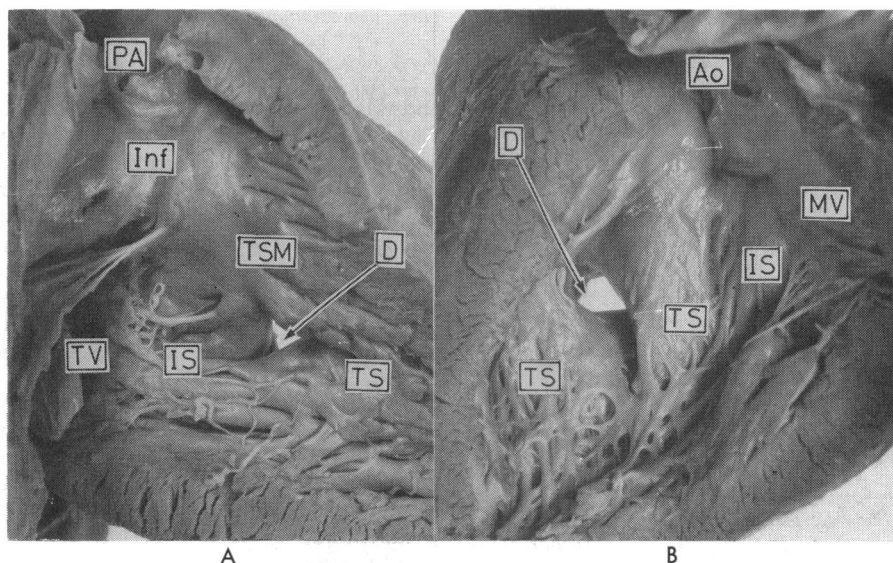


Fig. 8 A trabecular septal defect viewed from the right ventricle (A), and the left ventricle (B).

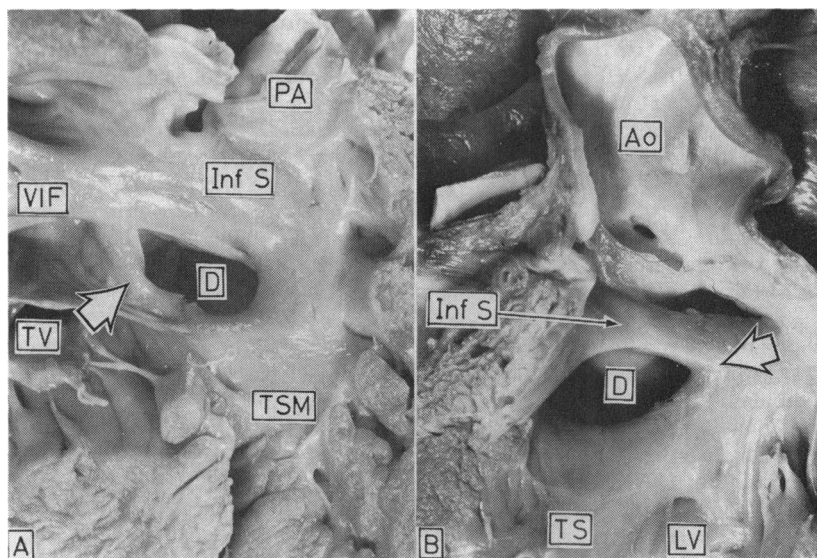


Fig. 9 A muscular defect of the infundibular septum viewed from the right ventricle (A), and the left ventricle (B). Note that the posterior limb of the trabecula septomarginalis (arrowed) fuses with the ventriculoinfundibular fold (VIF) in the posterior rim of the defect. The infundibular septum separates the defect from the pulmonary valve. The membranous septum is intact (abbreviations as before).

III: Subarterial infundibular defects

These defects were similar to the muscular infundibular defects except that the infundibular septum was totally deficient, so that the aortic and pulmonary valves were contiguous in the roof of the defect (compare Fig. 9 and 10). Because of this, the left coronary and right coronary aortic valve cusps

were free to move into the right ventricular outflow tract, and some of the hearts had aortic cusp prolapse. In the floor of the defect, as in the muscular infundibular defects, the posterior limb of the trabecula septomarginalis extended to the ventriculoinfundibular fold, forming a muscular rim in front of a normally formed membranous septum

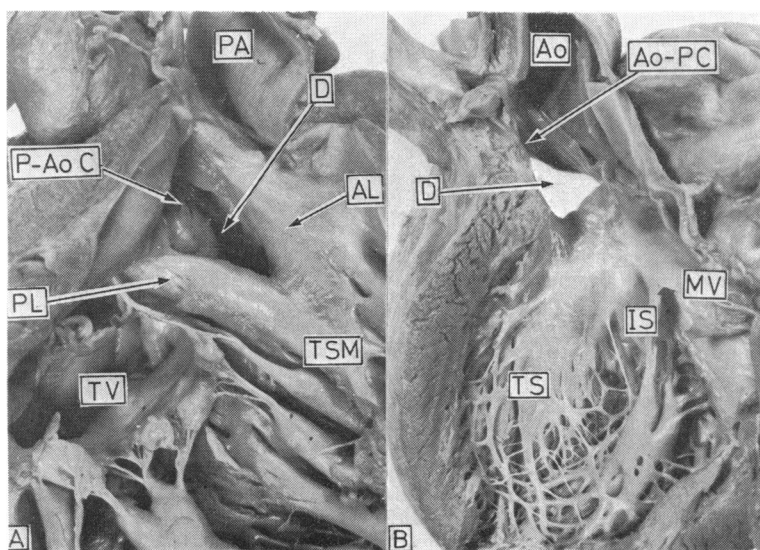


Fig. 10 Right ventricular (A) and left ventricular (B) views to a subarterial septal defect. The posterior limb (PL) of the trabecula septomarginalis fuses with the ventriculoinfundibular fold separating the defect from the normally formed membranous septum. Deficiency of the infundibular septum permits pulmonary to aortic fibrous continuity, P-AoC, Ao-PC.

(Fig. 10). When viewed from the left ventricle, the right and left coronary cusps of the aortic valve formed the entire roof of the defect, being contiguous with the pulmonary valve and distinguishing the defects from muscular infundibular defects (compare Fig. 9B and 10B).

ANGIOGRAPHIC CORRELATIONS

Examination of angiograms from patients studied at the University of Alabama in Birmingham (courtesy of Dr L M Barger Jr) showed that using projections designed to profile the ventricular septum⁷ it was possible to distinguish perimembranous (Fig. 11) from muscular (Fig. 12) and subarterial infundibular (Fig. 13) defects.⁸ Furthermore, careful assessment of each type showed that it was possible to distinguish the three varieties of both perimembranous (Fig. 11A-C) and muscular (Fig. 12A-C) defects.

Discussion

The classification of isolated ventricular septal defect here presented is an eclectic modification of categorisations of earlier workers⁵⁻⁹⁻¹⁴ together with our own earlier attempts to achieve a satisfactory nosology.^{2,3} In the present categorisation we have used purely descriptive terms to describe the three components of the muscular septum delineated on developmental grounds by Goor *et al.*,¹⁴ and we believe that this usage has increased the value of the categorisation. Certainly by using angiographic views designed to profile the ventricular septum⁷ it has proved feasible to distinguish defects existing in relation to these three parts of the muscular septum in addition to distinguishing between perimembranous, muscular, and subarterial defects.

Although it clearly has value in angiographic diagnosis, it is our contention that it has con-

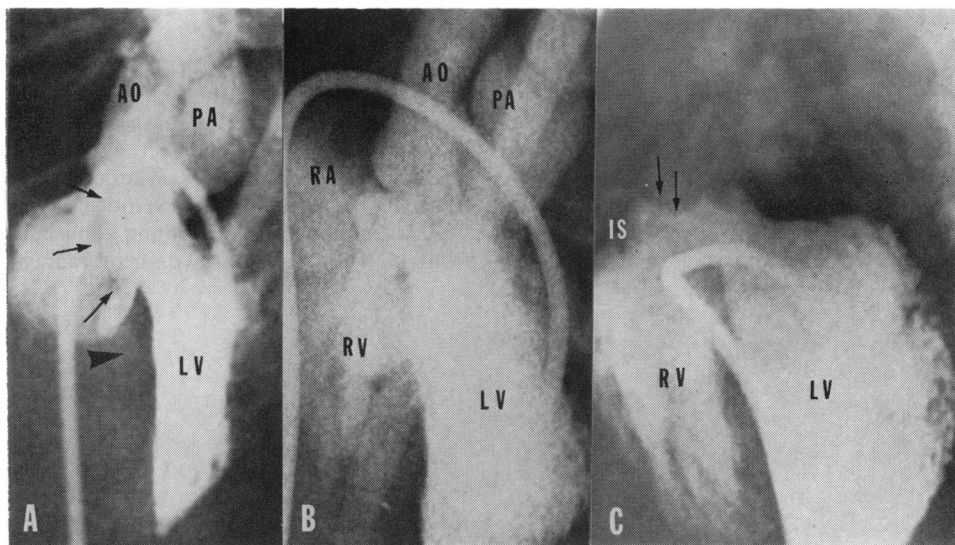


Fig. 11 Angiographic feature of perimembranous ventricular septal defects seen in 'four chamber' views. The diagnoses were subsequently verified at operation. (A) Perimembranous ventricular septal defect excavated in the inlet septum. The ventricular septal defect is large, roofed by the aortic valve. The inferior border is near the crux of the heart (arrow-head). A large segment of the septal tricuspid leaflet (arrow) is in contact with the defect, but both atrioventricular valves are well formed. AO, aorta; PA, pulmonary artery; LV, left ventricle. (B) Perimembranous defect excavated in the trabecular septum. The defect is roofed by the aortic valve (non-coronary cusp) and its anterior border is the upper portion of the trabecular septum (arrow). Notice that the contrast media from the left ventricle (LV) opacifies the right ventricle (RV) and also the right atrium (RA). The left ventricle—right atrium connection is through the medial commissure of the tricuspid valve which covers the ventricular septal defect. AO, aorta; PA, pulmonary artery. (C) Perimembranous defect excavated in the infundibular septum. The defect is located immediately beneath the right and non-coronary (aortic) cusps (arrows). The contrast media injected into the left ventricle (LV) passes into the right ventricle (RV) opacifying the trabeculated portion mainly. The infundibular septum (IS) is deviated anteriorly, leaving the right and non-coronary cusps above the right ventricle.

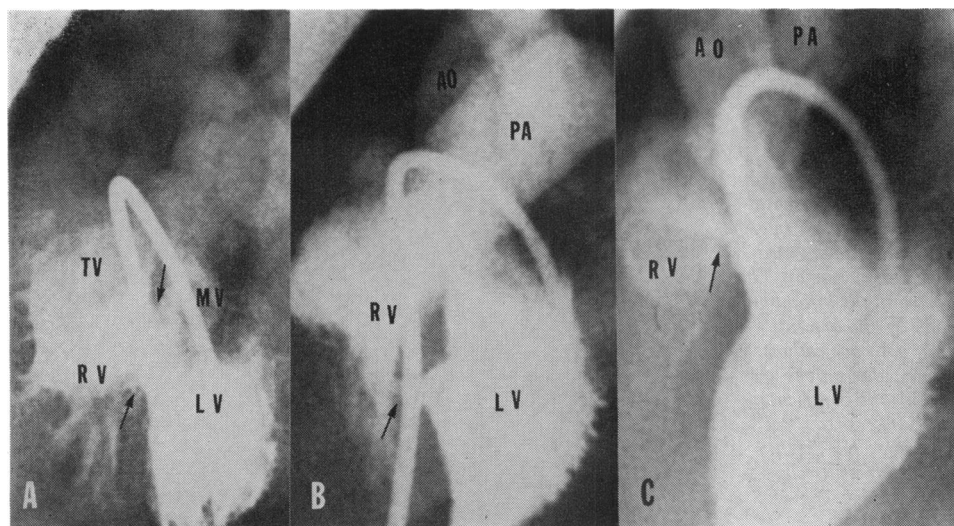


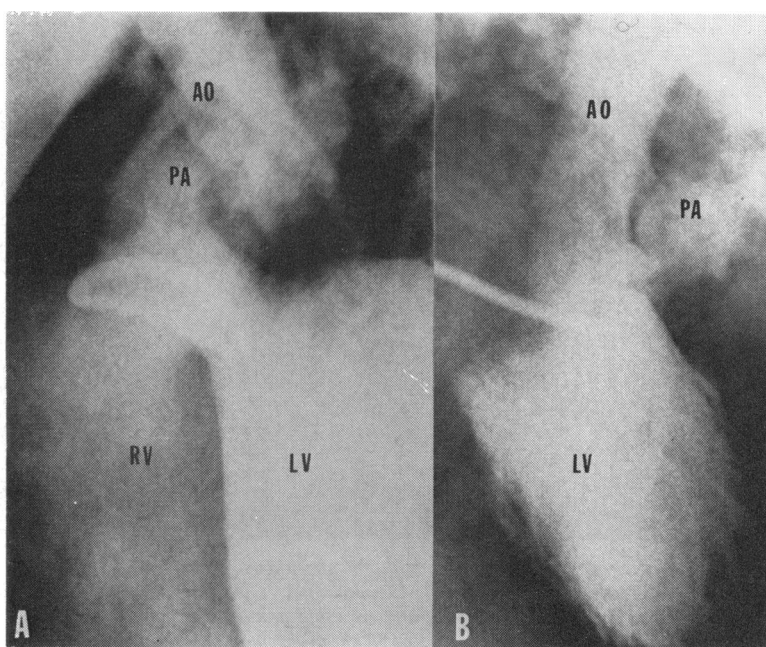
Fig. 12 Angiographic feature of muscular ventricular septal defects seen in 'four chamber' views. The diagnosis in each case was verified at surgery. (A) *Muscular inlet.* The defect (arrows) is located in the inlet portion of the ventricular septum discontinuous from the mitral and tricuspid annuli. RV, right ventricle; LV, left ventricle. (B) *Muscular trabecular.* The defect (arrow) is located in the trabecular portion of the septum near its posterior border. RV, right ventricle; LV, left ventricle; AO, aorta; PA, pulmonary artery. (C) *Muscular infundibular.* The defect (arrow) is in the right border of the left ventricular outflow tract, separated from the arterial and atrioventricular valves. Note the early opacification of pulmonary artery (PA). AO, aorta; RV, right ventricle; LV, left ventricle.

siderable value in surgical viewpoint. The main thrust of the surgical classification of Kirklin and his colleagues^{11 12} was to distinguish those defects in relation to the aortic valve ('high' defects) from those not in relation to the aortic valve ('low' defects). Our classification refines this possibility, since it distinguishes defects with entirely muscular rims, which clearly are never directly related to the aortic valve, from those with rims formed by intrinsically fibrous tissue in which the aortic valve forms part of this rim. It further distinguishes the latter group into those in which the aortic valve is related to the defect as part of the central fibrous body (perimembranous defects) from those in which it is related to the defect in continuity with the aortic and pulmonary valves (subarterial infundibular defects). However, the surgical value of the present classification goes far beyond the relation of the defect to the aortic valve. The recognition of a defect as perimembranous, muscular, or infundibular immediately alerts the surgeon to the likely disposition of the conduction tissues. If a defect is perimembranous, previous studies¹⁵⁻¹⁷ have shown that the atrioventricular conduction axis will always be related to the posteroinferior quadrant of the defect (to the surgeon's right hand as viewed from

either the right atrium (Fig. 14) or a right ventricular infundibulotomy). In contrast, a muscular defect in the inlet septum will have the conduction tissue related to its superoanterior quadrant (to the surgeon's left hand as viewed from atrium (Fig. 14) or infundibulum). Muscular defects in the trabecular septum are unlikely to be related to the non-branching or branching components of the conduction tissue axis, though they may be related to bundle-branches,¹⁷ while infundibular, muscular, or subarterial defects are unrelated to the conduction tissue.¹⁸ The recognition of a perimembranous defect as inlet, trabecular, or infundibular also provides information regarding the direct relation of the ventricular conduction tissue axis to the septal rim.¹⁸

In addition to providing this information which we believe to be of value to both diagnostician and surgeon, our study helped to clarify several points which previously had given us some problems. Firstly, it became evident that though most defects were in the region of the membranous septum, they represented more than mere absence of the membranous septum. Becu *et al.*⁵ have previously emphasised this point. Goor and his colleagues^{14 19} similarly recognised that most defects in this area

Fig. 13 *Angiographic appearance of subarterial ventricular septal defect, left ventriculogram in (A) 'long axial', and (B) elongated right anterior oblique views. The defect is located in the infundibular septum roofed by the arterial valves seen in both projections. The atrioventricular valves are not related with the ventricular septal defect. AO, aorta; PA, pulmonary artery; RV, right ventricle; LV, left ventricle.*



were not 'true' defects of the membranous septum, but they did recognise a small group which they categorised as membranous defects. In our series, we did not encounter any defects so small that we considered them truly to be mere absence of the interventricular membranous septum. Instead, it was our interpretation that all defects resulted from deficiency of the muscular septum in the environs of the interventricular membranous septum, which was frequently present as part of the border of the defects. Hence, our suggestion of the term 'perimembranous' to describe these defects, and our subdivision of the group depending on whether the muscular deficiency affected primarily the inlet, trabecular, or outlet components of the muscular septum. It is possible that tiny defects could result simply from absence of the interventricular membranous septum, or that alternatively a small perforation could be found in an aneurysmal membranous septum which has closed an existing defect. If required, the term 'membranous septal defect' could accurately be applied to such lesions.

The second point relates to the so-called 'isolated ventricular septal defect of persistent common atrioventricular type'.²⁰ As one of us has indicated, most of these defects are really defects of the inlet septum extending into the area of the membranous septum, and have presently been categorised as perimembranous inlet defects.² It was significant that not all the perimembranous inlet defects we

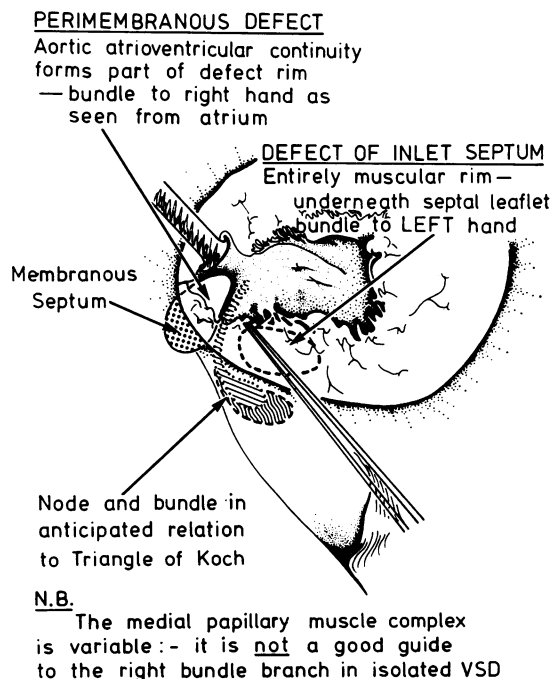


Fig. 14 *Diagrammatic representation of the different relations of perimembranous and muscular inlet defects to the atrioventricular conduction tissue axis as viewed by the surgeon from the right atrium.*

studied exhibited the stigmata of atrioventricular defects, namely disproportion between the inlet and outlet dimensions of the muscular septum and malorientation of the aortic valve. Indeed, not all the defects illustrated by Neufeld *et al.*²⁰ showed these features. Only four of the 55 inlet perimembranous defects we examined exhibited the 'scooped-out' septum so typical of the atrioventricular defect, two of these having clefts in their mitral valves. If the term 'isolated atrioventricular canal defect' is to be used, it is our preference to restrict it to these hearts, while still preferring the description 'perimembranous inlet defect with gross deficiency of the inlet septum' as being less ambiguous.

Thus, we have presented an eclectic classification of ventricular septal defects based on the premise that the ventricular septum has membranous and muscular portions, the latter itself having inlet, trabecular, and outlet components. We have shown that the classification, based on study of pathological material, is of value in the angiographic laboratory, and that the diagnostic precision thus afforded has considerable surgical significance. We offer the classification as a simple alternative to those presently available.

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